Complete Summary

GUIDELINE TITLE

Celiac disease.

BIBLIOGRAPHIC SOURCE(S)

National Institutes of Health (NIH) Consensus Development Panel on Celiac Disease. Celiac disease. Bethesda (MD): U.S. Department of Health and Human Services (DHHS); 2004 Aug 9. 15 p.

GUIDELINE STATUS

This is the current release of the guideline.

COMPLETE SUMMARY CONTENT

SCOPE

DISEASE/CONDITION(S)

Celiac disease

GUIDELINE CATEGORY

Counseling
Diagnosis
Evaluation
Management
Risk Assessment
Treatment

CLINICAL SPECIALTY

Allergy and Immunology
Endocrinology
Family Practice
Gastroenterology
Internal Medicine
Nutrition
Oncology
Pathology
Pediatrics
Preventive Medicine

INTENDED USERS

Dietitians
Health Care Providers
Nurses
Physicians

GUIDELINE OBJECTIVE(S)

- To improve awareness, diagnosis, and management of celiac disease
- To examine the current state of knowledge regarding celiac disease and to identify directions for future research. Specifically, the following key questions were addressed:
    - How is celiac disease diagnosed?
    - How prevalent is celiac disease?
    - What are the manifestations and long-term consequences of celiac disease?
    - Who should be tested for celiac disease?
    - What is the management of celiac disease?
    - What are the recommendations for future research on celiac disease and related conditions?

TARGET POPULATION

Patients with confirmed or suspected celiac disease

INTERVENTIONS AND PRACTICES CONSIDERED

Diagnosis and Risk Assessment

1. Diagnostic Tests
    - Immunoglobulin A (IgA) antihuman tissue transglutaminase (TTG) test
    - IgA endomysial antibody immunofluorescence (EMA) test
    - Biopsies of the proximal small bowel in individuals with a positive celiac disease antibody test, except those with biopsy-proven dermatitis herpetiformis.
2. Standardization of pathology reports following biopsy of the small bowel
3. Management of patients with suggestive symptoms and a negative serology test (e.g., additional testing, trial of gluten-free diet)
4. Role of genetic marker testing (human leukocyte antigen [HLA] haplotypes)

5. Risk assessment
6. Identification of individuals who should be tested for celiac disease

Management of Celiac Disease

1. Consultation with a dietitian
2. Patient education regarding celiac disease
3. Lifelong adherence to a gluten free diet
4. Identification and treatment of nutritional deficiencies
5. Access to an advocacy group
6. Continuous long-term follow-up by a multidisciplinary team
7. Screening for osteoporosis
8. Treatment of complications associated with celiac disease

MAJOR OUTCOMES CONSIDERED

- Sensitivity and specificity of serological tests for celiac disease
- Value of standardized pathology criteria for the diagnosis of celiac disease
- Symptom resolution following adoption of a gluten-free diet
- Value of genetic marker testing (human leukocyte antigen [HLA] haplotypes) in the diagnosis of celiac disease
- Prevalence rates of celiac disease and identification of high risk groups who may benefit from screening
- Disease manifestations, long-term consequences, and complications
- Morbidity and mortality associated with celiac disease

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

The Agency for Healthcare Research and Quality (AHRQ) supported the National Institutes of Health (NIH) Consensus Development Conference on Celiac Disease through its Evidence-based Practice Center (EPC) program. Under contract to the AHRQ, the University of Ottawa EPC developed the systematic review and analysis that served as a reference for discussion at the conference. The National Library of Medicine in collaboration with the University of Ottawa EPC conducted the literature search.

A series of systematic reviews on five areas of celiac disease (CD) were completed:

1. Sensitivity and specificity of serological tests
2. Prevalence and incidence of CD
3. CD-associated lymphoma
4. Consequences of testing for CD
5. Interventions for the promotion and monitoring of adherence to a gluten-free diet (GFD)

Staff at the National Library of Medicine performed a series of searches in support of the literature review of celiac disease. Searches were run in the MEDLINE® (1966 to Oct 2003) and EMBASE (1974 to Dec 2003) databases for each of the five objectives and their respective sub-objectives separately. Furthermore, for the 4th and 5th objectives, PsycINFO (1840 forward), AGRICOLA (1970 forward), CAB (1972 forward), and Sociological Abstracts (1963 forward) database searches were run in December 2003.

Study selection for each objective was performed using three levels of screening with predetermined increasingly more strict criteria to ensure that all relevant articles were captured. Following a calibration exercise, two reviewers independently screened all studies using a Web-based system that allowed automatic identification of review disagreements. These disagreements were resolved by consensus.

For each CD objective, a detailed and standardized data abstraction form was developed. For each objective, data abstraction was conducted by one reviewer and verified by another. The extracted data was further verified by one of the principal investigators. Quality assessments were performed using specific instruments for each of the included study types.

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Expert Consensus

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Not applicable

METHODS USED TO ANALYZE THE EVIDENCE

Systematic Review

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

The data obtained from this review fell into several broad categories, which correspond in large part to the individual study objectives. Data for the sensitivity and specificity of each serological marker was considered separately, and studies were further divided according to the age group of the study population. Attempts were made to identify, explain, and minimize clinical and statistical heterogeneity in the included studies. A Pearson's Chi Square with n-1 degrees of freedom, where n represents the number of included studies in an analysis, was calculated to assess statistical heterogeneity. Pooled estimates were only calculated, if clinically and statistically appropriate. In situations where pooling was not performed, a qualitative systematic review was conducted.

To produce clinically useful pooled statistics, a weighted mean of the overall sensitivity and specificity from the included studies was calculated, along with 95-percent confidence intervals (CIs). The pooled estimates for the sensitivity and specificity were compared with a summary receiver operating characteristic (ROC) curve, calculated for the same group of studies as a second check of the estimates.

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Expert Consensus (Consensus Development Conference)

DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

The National Institutes of Health (NIH) convened a Consensus Development Conference on Celiac Disease on June 28–30, 2004. The National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK) and the Office of Medical Applications of Research (OMAR) of the NIH were the primary sponsors of this meeting. The U.S. Food and Drug Administration, the U.S. Department of Agriculture, the National Institute of Child Health and Human Development, the National Cancer Institute, and the National Institute of Allergy and Infectious Diseases were the cosponsors.

This two-and-a-half-day conference examined the current state of knowledge regarding celiac disease and identified directions for future research. During the first day-and-a-half of the conference, experts presented the latest celiac disease research findings to an independent panel. After weighing this scientific evidence, the panel drafted a statement that addressed the key questions.

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Not applicable

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

On the final day of the conference, the panel chairperson read the draft statement to the conference audience and invited comments and questions.

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

Conclusions

Celiac disease is an immune-mediated intestinal disorder with protean manifestations. Celiac disease is common, affecting 0.5 to 1.0 percent of the general population of the United States, but is greatly underdiagnosed. There are now specific and sensitive serologic tests available to aid in diagnosis that need to be more widely applied. The treatment of celiac disease remains a lifelong gluten-free diet, which results in remission for most individuals. The classic presentation of diarrhea and malabsorption is less common, and atypical and silent presentations are increasing. Most individuals are being seen by primary care providers and a broad range of specialists. Therefore, heightened awareness of this disease is imperative. Education of physicians, registered dietitians, and other health providers is needed.

The panel recommends the following:

- Education of physicians, dietitians, nurses, and the public about celiac disease by a trans-National Institutes of Health (NIH) initiative, to be led by the National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK), in association with the Centers for Disease Control and Prevention.
- Standardization of serologic tests and pathologic criteria for the diagnosis of celiac disease.
- Adoption of a standard definition of a gluten-free diet based on objective evidence such as that being developed by the American Dietetic Association.
- Development of an adequate testing procedure for gluten in foods and definition of standards for gluten-free foods in the United States to lay the foundation for rational food labeling.
- Formation of a federation of celiac disease societies, celiac disease interest groups, individuals with celiac disease and their families, physicians, dietitians, and other health care providers for the advancement of education, research, and advocacy for individuals with celiac disease.

CLINICAL ALGORITHM(S)

None provided

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The panel, answering predefined questions, developed their conclusions based on the scientific evidence presented in open forum and the scientific literature.

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

Improved awareness, diagnosis, and management of celiac disease

POTENTIAL HARMS

False positive or false negative serological test requiring additional investigative procedures

QUALIFYING STATEMENTS

QUALIFYING STATEMENTS

- The statement reflects the panel´s assessment of medical knowledge available at the time the statement was written. Thus, it provides a "snapshot in time" of the state of knowledge on the conference topic. When reading the statement, keep in mind that new knowledge is inevitably accumulating through medical research.
- This statement is an independent report of the panel and is not a policy statement of the NIH or the Federal Government

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Living with Illness

IOM DOMAIN

Effectiveness
Patient-centeredness

IDENTIFYING INFORMATION AND AVAILABILITY

BIBLIOGRAPHIC SOURCE(S)

National Institutes of Health (NIH) Consensus Development Panel on Celiac Disease. Celiac disease. Bethesda (MD): U.S. Department of Health and Human Services (DHHS); 2004 Aug 9. 15 p.

ADAPTATION

Not applicable: The guideline was not adapted from another source.

DATE RELEASED

2004 Aug 9

GUIDELINE DEVELOPER(S)

National Institutes of Health (NIH) Consensus Development Panel on Celiac Disease - Independent Expert Panel
Office of Medical Applications of Research (NIH) - Federal Government Agency [U.S.]

GUIDELINE DEVELOPER COMMENT

National Institutes of Health (NIH) consensus and state-of-the-science statements are prepared by independent panels of health professionals and public representatives on the basis of (1) the results of a systematic literature review prepared under contract with the Agency for Healthcare Research and Quality (AHRQ), (2) presentations by investigators working in areas relevant to the conference questions during a 2-day public session, (3) questions and statements from conference attendees during open discussion periods that are part of the public session, and (4) closed deliberations by the panel during the remainder of the second day and morning of the third. This statement is an independent report of the panel and is not a policy statement of the NIH or the Federal Government.

SOURCE(S) OF FUNDING

United States Government

GUIDELINE COMMITTEE

National Institutes of Health (NIH) Consensus Development Panel

COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

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FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

Not stated

GUIDELINE STATUS

This is the current release of the guideline.

GUIDELINE AVAILABILITY

Electronic copies: Available in Portable Document Format from the National Institutes of Health (NIH) Consensus Development Conference Program Web site.

Print copies: Available from the NIH Consensus Development Program Information Center, PO Box 2577, Kensington, MD 20891; Toll free phone (in U.S.), 1-888-NIH-CONSENSUS (1-888-644-2667); autofax (in U.S.), 1-888-NIH-CONSENSUS (1-888-644-2667); e-mail: consensus_statements@mail.nih.gov.

AVAILABILITY OF COMPANION DOCUMENTS

The following is available:

- Rostom A, Dubé C, Cranney A, et al. Celiac Disease. Summary, Evidence Report/Technology Assessment: Number 104. Rockville (MD): Agency for Healthcare Research and Quality; AHRQ Publication Number 04-E029-1; 2004 Jun.

Electronic copies available from the Agency for Healthcare Research and Quality (AHRQ) Web site.

PATIENT RESOURCES

None available

NGC STATUS

This NGC summary was completed by ECRI on October 7, 2004. The information was verified by the guideline developer on November 16, 2004.

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